

Figure 2. This section shows tumour invading the infraorbital nerve ($H\&E, \times 250$)

and with the desmoplastic stroma at the tumours edge. This feature can lead to serious difficulty for the pathologist, especially if the surgeon is relying on the results of frozen section to assess tumour clearance.

Major differential diagnoses are distinguished by stains, immunohistochemistry and electron microscopy. Immunohistochemical use of keratin markers and leucocyte common antigen will detect the carcinomas and lymphomas. However, S100 positivity which is found mainly in neural crest derived tumours will therefore not distinguish between malignant schwannoma and malignant melanoma.

The similarity between these two tumours was initially reported by Reed and Leonard⁸, who described a variant of desmoplastic melanoma which shared certain histological features with malignant epithelioid schwannoma. Their case, however, involved a pigmented lesion arising in an area of lentigo. Enzinger and Weiss further clarified the picture when, in 1983 they described a 'malignant epithelioid schwannoma of soft tissue origin'. They too recognized the similarity between this tumour and malignant melanoma but defined it as a schwannoma on the basis of two key features; firstly the absence of melanocyte activity within the epidermo-dermal junction and secondly the absence of melanin pigment¹.

Electron microscopy has revealed other features to aid distinction including a distinct basal lamina. The most important however, is the absence of melanosomes or pre-melanosomes characteristic of melanoma⁹.

The natural history of those tumours arising below the clavicles suggests a virulent tumour. In a study of 14 patients nine died, six from metastatic disease within 2 years

of diagnosis. Spread occurred via both haematogenous and lymphatic routes with the low incidence of local recurrence being attributed to radical excision of the primary lesion⁴.

As indicated reports of tumours arising on the head and neck are few and one has to be guarded in characterizing its behaviour. Of three reported cases, two died of metastatic disease 3 and 6 years after presentation^{5,7}. Significantly both patients had prolonged problems with locally recurrent disease necessitating repeated attempts at excision prior to rapid spread of the tumour and death. In the case reported by Bleach⁵, radical surgery was not carried out initially due to the patient's wishes, however repeated excision resulted in considerable morbidity. Radiotherapy did not provide any additional benefit. Chemotherapy was given in Chu's report⁷, but only at an advanced stage (the author does not give any details of the regimen used). The third case was well at 3 years follow-up with no evidence of spread6. However, in the light of experience of both our case and those above, one may debate as to whether the patient was diseasefree. Furthermore, prolonged follow-up would be essential. Experience from those cases discussed suggests that radical excision is indicated to achieve local control. Follow up will then be required over a 5 year period or longer.

References

- 1 Enzinger FM, Weiss SW. Soft tissue tumours. St Louis: CV Mosby, 1983:625-56
- 2 Daimaru Y, Hashimoto H, Enjoji M. Malignant "Triton" tumours: a clinico-pathologic and immunohistochemical study of nine cases. Hum Pathol 1984;15:768-78
- 3 Woodruff JM, Chernick NL, Smith MC, Millett WB, Foote FW. Peripheral nerve tumours with rhabdomyosarcomatous differentiation. Cancer 1973:32:426-39
- 4 Lodding P, Kindblom LG, Angervall L. Epithelioid malignant schwannoma, a study of 14 cases. Virchows Arch 1986;409:433-51
- 5 Bleach NR, Keen CE, Dixon JA. Superficial malignant schwannoma on the face: a case for early radical surgery. J Laryngol Otol 1989;103:316-18
- 6 Morgan K, Gray C. Malignant epithelioid schwannoma of soft tissue? A case report with immunohistology and electron microscopy. *Histopathology* 1985;9:765-75
- 7 Chu T, Shmookler BM. Malignant epithelioid schwannoma: a light microscopic and immunohistochemical study. J Surg Oncol 1988;39:68-72
- 8 Reed RJ, Leonard DD. Neurotropic melanoma. A variant of desmoplastic melanoma. Am J Surg Pathol 1979;3:301-11
- 9 Taxy JB, Battifora H, Trujillo Y, Dorfman HD. Electron microscopy in the diagnosis of malignant epithelioid schwannoma. Cancer 1981;48:1381-91

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CMV colitis in an immunocompetent adult

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Mild cytomegalovirus (CMV) infection is common, but very rarely causes colitis in immunocompetent adults. We report the first UK case of CMV colitis in the immunocompetent. The patient, the fourth case in the world literature to survive, is the first immunocompetent individual in whom the antiviral agent foscarnet has been used therapeutically.

Case report

A 35-year-old previously fit man presented with a 9 day history of increasing diarrhoea, malaise and nightsweats. Loose stools of small volume, with mucus but no blood were passed up to 12 times a day.

On examination, he was unwell but not dehydrated, with a pyrexia of 39.5°C. Abdominal examination including sigmoidoscopy was normal. An initial diagnosis of infective diarrhoea was made, and he was started on codeine, ampicillin and metronidazole. Stool cultures remained negative and colonoscopy was performed. This showed five large almost circumferential areas of superficial ulceration between the sigmoid and hepatic flexure. There was minimal reaction at the edges and no nodularity; the rest of the mucosa was endoscopically normal. Histology of the ulcers showed granulation tissue including scattered large cells with intranuclear and cytoplasmic inclusions which stained for cytomegalovirus antigen by the immunoperoxidase technique. There was otherwise no focal active inflammation

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0141-0768/92/ 040238-02/\$02.00/0 © 1992 The Royal Society of Medicine with normal lymphocytes and plasma cells and no evidence of long-standing colitis.

CMV serology showed a rising CFT titre and was positive for both IgG and IgM antibodies. HIV serology was negative, and repeated lymphocyte subsets were all normal. There was a mild hepatitis with alkaline phosphatase 409 u/l (normal 90-250) and AST 130 u/l (normal 10-55). Clinically, there was a small area of retinitis in one eye. CT scan of thorax and abdomen showed no evidence of malignancy. Bone marrow aspirate was normal.

The patient had a continued swinging pyrexia and persistent diarrhoea leading to a falling serum albumin of 22 g/l and Hb of 8.8 g/dl. In view of this, the antiviral agent foscarnet was commenced by intravenous injection in a dose of 12 g twice daily. The temperature gradually returned to normal after 12 days treatment, and diarrhoea reducing to four times per day. Treatment was continued for three weeks. Two months later, the retinitis and hepatitis had resolved. Repeat colonoscopy showed some persistent superficial ulceration, but other lesions had resolved. A reducing course of oral mesalazine was therefore given for 2 months until there was a complete resolution of all symptoms. There were no further symptoms at one year follow-up.

Discussion

CMV infection is common in immunocompetent people, causing a mild flu-like illness. Up to 81% of normal healthy individuals over the age of 35 have raised CMV titres^{1,2}. However, severe infections causing colitis are rare and usually fatal, although three surviving cases have been reported in the USA³⁻⁵. In contrast, CMV colitis is a common opportunistic infection in immunosuppressed individuals, principally in those who are HIV seropositive⁶⁻⁸. The reason for CMV colitis in immunocompetent subjects is unclear.

The three surviving cases reported in Washington included one male and one female whose symptoms were mild and resolved spontaneously within 4 weeks. The third case was an elderly woman with a clinical diagnosis of colonic Crohn's disease treated with steroids and sulphasalazine; however, histology showed CMV inclusions in granulation tissue. She made a good clinical recovery, within 6 months and no recurrence in 6 years.

The current case is different in several respects. The patient had never had any rectal trauma, and the severity of the illness was much more marked, with retinitis and hepatitis in addition to the colitis. In the light of the severity of the illness and the failure of symptoms to resolve spontaneously, the new anti-CMV agent, foscarnet, was given for the first time to an immunocompetent patient.

While CMV may be an incidental histological finding in many tissues, here it was only present at the sites of acute inflammation. There was a rising CMV titre associated with the illness which resolved in response to a specific anti-CMV drug (foscarnet). We conclude that this was a colitis caused by CMV, and despite extensive tests, there was no evidence of immunosuppression. The reason why the patient developed CMV colitis will remain an enigma. However, the long-term prognosis should be good as none of those who have survived CMV colitis (whether immunocompetent⁵, or immunosuppressed with drugs⁶) have had any long-term sequelae.

References

- 1 Weller TH. The cytomegaloviruses; ubiquitous agents with protean clinical manifestations. N Engl J Med 1971;285:203-14, 267-74
- Wright HT Jr. Cytomegaloviruses. In: Kaplan AS, ed. The herpes viruses New York: Academic Press, 1973:353-88
- 3 Wolfe BM, Cherry JD. Hemorrhage from cecal ulcers of cytomegalovirus infection. Am Surg 1973;177:490-4
- 4 Tamura H. Acute ulcerative colitis associated with cytomegalic inclusion virus. Arch Pathol 1973;96:164-7
- 5 Surawicz CM, Myerson D. Self-limited cytomegalovirus colitis in immunocompetent individuals. Gastroenterology 1988;94:194-9
- 6 Sidi S, Graham JH, Razvi SA, Banks PA. Cytomegalovirus infection of the colon associated with ulcerative colitis. Arch Surg 1979;114:857-9
- 7 Frank D, Raicht RF. Intestinal perforation associated with cytomegalovirus infection in patients with acquired immunodeficiency syndrome. Am J Gastroenterol 1983;78:167-9
- 8 McDonald GB, Shulman HM, Sullivan KM, Spencer GD. Intestinal and hepatic complications of human bone marrow transplantation. Part III. Gastroenterology 1986;90:770-84

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Association of migrainous headaches with Gasserian ganglial ablation

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Both the vascular and the neurogenic hypotheses are considered to be of fundamental importance in the pathogenesis of migraine. The two concepts are not incompatible¹, as neurogenic release of vasoactive substances via the facial and trigeminal nerves may cause painful vasodilatation sensed by the trigeminal nerve.

A direct link between the trigeminal nerve and migraine does not appear to have been reported. We present a patient in whom migrainous headaches developed following Gasserian ganglion ablation.

Case report

A 77-year-old man presented with severe unilateral headaches. Two years previously he had developed trigeminal neuralgia that was resistant to carbamazepine and had required Gasserian ganglial ablation later that year. Since then he had experienced ipsilateral headaches of increasing severity and frequency. The intercephalalgic gap progressively shortened until he was only achieving a few symptom-free hours.

These headaches last for 15-30 min. An aura occurred 20 min before the headache: numbness in the supraorbital region was followed by visible erythema of the frontal region, extending to the vertex. The headache was principally unilateral, though rarely bilateral, but did not extend posterior to the vertex. It was markedly distressing and accompanied by nasal stuffiness, hyperacusis and photophobia. His blood pressure rose markedly and his temperature slightly during headaches. He became increasingly confused and ultimately developed a right sided hemiparesis at the time of a headache.

He was admitted for investigation and treatment. Dental assessment was normal. CT scanning demonstrated an increase in CSF spaces but no focal abnormality. Chest X-ray and electrocardiography were consistent with left ventricular hypertrophy. Normal results were returned for the following investigations: full blood count, erythrocyte

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